Neonatal Cardiac Surgery [20]

JOSÉ FRAGATA
Department Cardiotoracic, Surgery Pediatric Unit
Hospital de Santa Marta, Lisboa, Portugal

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ABSTRACT

Key words
Surgery; Neonates; Post-operative care

RESUMO

Cirurgia Cardíaca Neonatal

Palavras-Chave
Cirurgia; Recém-nascidos; Cuidados pós-operativos

INTRODUCTION

Over the last twenty years neonatal cardiac surgery has gradually expanded in both figures and case complexity, complete corrections for neonates now being achieved more and more often.

In Europe, at the present, neonates represent 16.7 % of all pediatric cardiac procedures, but this particular patient group remains difficult to deal with if one considers the over-all mortality of 17.7 % – around three times the mortality observed for all pediatric patients (5.5 %) and also the mean length of stay in hospital, which is 15.9 days for neonates and 11 days for all patients [1].

Neonates are of smaller size and less weight than infants and children, but they also have immature organs and systems, tolerating surgical aggressions and particularly cardiopulmonary bypass poorly. Neonatal cardiac surgery needs surgical expertise as much as optimal post-operative care, in units with dedicated facilities for neonates, and only with a combination of both can good results be achieved.

Surgical options for neonates are divided between palliation or correction of the cardiac defect and, over the last ten years, the pressure for early correction now seems totally justified. However, one should not ignore the fact that results depend on the experience of both the center and the surgeon, and that the case load might play a role here [2]. Finally, immediate results are not all that counts; long-term results matter as well, particularly fine neurological function, which has been shown to be affected by extended periods of total circulatory arrest, a method that is still used for very complex corrections, notably of the aortic arch [3].

The immediate surgical risk, together with the expected long-term outcome are both to be balanced with the possible known sequelae of the cardiac lesion itself on the heart (volume or pressure load), on the lungs and pulmonary circulation (pulmonary artery disease), and on the baby's overall development (congestive heart failure, cyanosis, etc.) and also the impact of the cardiopathy on the baby's family, before any decisions concerning treatment options are made.

It is clear that strong pressure on early correction seems more than desirable, provided that benchmarked results are favorable.

Perioperative care for neonates

- *Care prior to the operation;*
- *Extra-corporeal circulation;*
- *Organ protection;*
- *Post-operative care.*

Neonates often need some sort of medical optimization prior to surgery [4]. For most elective procedures the operation should be

conducted only after the first week of life, when most of the initial neonatal adaptations to extra-uterine life have taken place.

Most diagnoses are now made non-invasively by echocardiography. Cardiac catheterization and angiography are less and less used in the critically ill neonate, as they seem to be detrimental, especially for renal function and the baby's general condition.

Babies with obstructive lesions to the left side of the heart, and particularly those with aortic arch obstruction, need immediate opening of the ductus arteriosus by prostaglandin E infusion (0.05-0.5 mcg/kg/m), mechanical ventilation, inotropic support and diuretics in order to restore peripheral systemic perfusion, support the heart, normalize blood gases, particularly arterial pH and lactate levels. Surgery for complex left-sided lesions should wait until the baby has been resuscitated, is stable and passing urine adequately, has normal pH and is metabolically corrected; for most babies this can be accomplished in not more than 24 to 48 hours. Nutrition, also, is of paramount importance for any neonate undergoing surgery, especially in the presence of congestive heart failure, due to the fact that body fat and carbohydrate supplies are limited in neonates and consumption is dramatically increased by surgical stress.

For babies with obstructive lesions to the right side combined with lung hypoperfusion, ductal opening might be the only way to perfuse the lungs, reverse cyanosis and tissue hypoxia and correct acidosis. These babies do much better off the ventilator – unless they are very sick – and should immediately be started on i.v. prostaglandin (PGE). Surgery shall take place only after adequate resuscitation is achieved.

Another group is complex situations such as TGA and TAPVD, where the problem is deficient mixing. These patients undergo emergent atrial septostomy to improve atrial mixing, improve circulation dynamics and fight progressive acidosis.

TAPVD, especially if obstructed, as is the rule in the neonate presenting with the infra-cardiac type, combines mixing defect with obstruction of venous drainage and represents a genuine surgical emergency. No time should be wasted trying to optimize these patients, other than ventilating them and sending them to surgery straight away.

Cardiopulmonary bypass (CPB) for neonates

Extra-corporeal circulation is poorly tolerated by neonates, due to the dilution effect of the priming volume, due to the pro-inflammatory component of CPB that is mediated by the interaction between the foreign surfaces of the circuit and blood, through complement cascade activation and neutrophil and platelet mediation [5]. Direct trauma to the blood cells, different flows and a broad range of temperatures, all contribute to post-pump syndrome characterized by capillary leak, pulmonary vascular resistance fluctuation and tendency to bleed, together with multiorgan sequelae. Target organs are the heart, lungs, kidneys and brain, where ventricular hemorrhage may occur, especially in prematures.

Some of these damaging effects may be minimized by appropriate CPB policy – small priming volumes, the use of heparin-coated systems, filters on the arterial line, vasodilatation during bypass with an α-blocker drug (phenoxybenzamine) [6] and the conduction of perfusion with high flow (150 ml/kg) and a low pressure regime, at all costs minimizing circulatory arrest periods. All these measures seem to be protective, promoting better tissue perfusion.

The alpha-stat policy, which lets pH drift with hypothermia and cooling, is both safe and practical in use, although it is still the subject of debate, as it would in theory restrict cerebral blood flow at low temperatures and extremely alkaline pH [7].

Even then, capillary leak does occur and extra vascular water tends to accumulate, particularly in the heart, lungs, brain and extra-cellular tissue. The shortest bypass times and the use of an appropriate CPB policy certainly attenuate capillary leakage. Modified ultrafiltration (MUF), introduced by the Great Ormond Street group, is now used routinely to remove extra fluid and to promote hemoconcentration after CPB in children and specially in neonates [8]. Its beneficial effects have been demonstrated on reduction of total body water, but also upon myocardial and lung function, as well as on the coagulation cascade. It seems that, besides its hemoconcentration action, some anti-inflammatory effect might be expected from the use of MUF, by filtering and removing inflammation mediators such as interleukins

1.6 and TNF-α, but this has yet to be proved [9, 10].

Achieving complete hemostasis is important for any cardiac operation, but it is crucial for neonates. Careful surgical technique, warming of the patients (clotting is not normal in hypothermic babies), adequate heparin reversal, aprotinin and selected blood components (FFP and platelets) are routinely used to achieve hemostasis. It is essential to remember that bleeding promotes more bleeding, due to local fibrinolysis, and also that tamponade is common in neonates that are bleeding. Surgical re-exploration should not be subject to too much thought or unnecessary delay in neonates.

Table I

Cardiopulmonary bypass protocol and organ protection for neonates

- Neonatal – Safe Micro oxygenator + mini-circuit – priming volume < 300 ml
- Priming: CPD blood* – 100 ml + FF plasma – 100 ml + NS – 100 ml + 20% albumin – 10 ml + NaHCO3 -1 mEq/kg + mannitol 20% - 25 ml + aprotinin - 20 000 units/kg + methylprednisolone 30 mg/kg
- Heparin – 300 units/kg for ACTs > 600 sec (check every 30 min)
- Flow at 37°C – 2.8 l/m/m²
  Vasodilatation – phenoxybenzamine 1 mg/kg
- Reperfusion protection – 20% mannitol - 1 ml/kg (unclamping) + Cagl 500 mg (at 36°C)
- Ultrafiltration: Modified ultrafiltration – 700 ml/m² filtrated fluid over 15-20 min

* less than 2 days old.

Organ protection

All the systems and organs in the neonate are at risk during the operation and whenever using CPB, due to known inflammatory reactions and, particularly because of immaturity – the heart, brain and kidneys, together with the coagulation cascade need individualized consideration [11]. Experience has indicated that the pre-operative use of steroids is protective to the cell membranes and, accordingly, methyl- prednisolone (30 mg/kg) is given prior to CPB. Aprotinin is claimed to spare platelets and to have some anti-inflammatory action and so 10.000 U/kg aprotinin is used for all cases, as a bolus at the beginning of the operation. Judicious hemodilution (20-30% hematocrit) is acceptable for both rheology, at low temperatures, and oxygen transport together with appropriate pump flows, and pharmacologically induced vasodilatation with regitine (1 mg/kg) all seem to reduce the damaging effects of CPB. Short bypass times and MUF are also critical in minimizing pump deleterious effects. The heart needs to be protected from ischemia by both cold and cardioplegia. The neonatal heart is resistant to ischemia but it is also sensitive to capillary leakage and edema accumulation. Moreover, the immature heart has low energy supplies, few myocytes and contractile elements and inefficient calcium removal mechanisms. As a consequence of all these immature characteristics, both systolic and dia-stolic failure are common after surgery, in what might be considered a stunning (ischemia/reperfusion) lesion. Keeping cross-clamping times as short as possible, using cardioplegia (crystalloid or blood – 10 ml/kg once only), at deep hypothermia (17-20°C), or every 30 min if temperature is above 25°C and, especially, giving it by gentle hand injection (infusion pressure < 30 mmHg), as well as preventing perfusion pressures above 50 mmHg, at all times, are simple protective measures for the neonatal heart.

The brain is at risk from emboli related to CPB, but particularly when circulatory arrest is used, as is still nowadays the case for most procedures on the aortic arch. The Boston Children's Hospital Circulatory Arrest Study has shown, as well as acute changes (seizures), neurodevelopmental abnormalities at one year of age when long (> 45 m) circulatory arrest periods were used in neonates, after the arterial switch operation [3].

The lungs also suffer from transient compliance deterioration and gas-exchange dysfunction, due to interstitial edema and Va/Q changes but, particularly, endothelial dysfunction due to leukocyte sequestration leads to insufficient NO production and favors pulmonary vascular tone fluctuation and, in some patients, such as neonates or in children with pulmonary vascular disease, prompts a pulmonary hypertensive crisis [12].

The incidence of bleeding due to hematological abnormalities is around 15%, and renal and hepatic failure after CPB in neonates are present in not more than 5% [13].

Myocardial failure is common after any cardiac operation. Typically, even patients doing well show around 40% decrease in LV systolic function and usually deteriorate

Table II

Post-operative care for neonates

- **Ventilator settings:**
  IPPV, tidal volume 10-15 ml/kg, rate 30/m, Insp time – 33%, FiO_2 . 75
- **Sedation:**
  Morphine sulfate: 0.1 mg/kg or 0.5 mg/kg in 50 ml D 5%: 1 to 6 ml/h
  Midazolam: 0.1 mg/kg or 3 mg/kg in 50 ml D 5%: 1 to 6 ml/h
- **Maintenance fluid:**
  Dx 10% 1/4 saline + {KCl 3 mEq + CaGluc 500 mg + $MgSO_4$ 0.4 mg} / 100 ml
  Post-op day 1 and 2 – 2.5 ml/kg/h, post-op day 3 and on – 4 ml/kg/h
- **Volume loading:**
  Ht >40% – 5% albumin/plasma 5-10 ml/kg
  Ht <40% - Blood – 5 ml /kg (+ 1 ml/kg CaGluc / 100 ml CPD colloid)
- **Inotropes:**
  Dopamine – 2 -10 mcg/kg/m + milrinone 50 mcg/kg load + 0.25-1 mcg/kg/m
  Dobutamine – 2 to 10 mcg/kg/m
  Adrenaline – 0.05 – 0.5 mcg/kg/m
- **Vasodilators:**
  Nitroglycerin – 0.5 – 10 mcg/kg/m
- **Diuretics:**
  Furosemide 1 mg / kg + aminophylline 3 mg/kg + albumin 20% 1.5 ml /kg
  Furosemide: 0.1 – 0.5 mg/kg/h, as perfusion
- **Renal failure:**
  fluid overload and rising potassium (K >5.5 mEq)
  Peritoneal dialysis: 20 ml / kg 30 m cycles alternating 1.5 /4.25% solutions
  K 6 mEq: 30% dextrose – 1.5 ml/kg + insulin 0.25 u/kg + CaGluc 50 mg/kg
- **Pulmonary HT:**
  Sedation – fentanyl 1 - 3 mcg/kg/m pancuronium 0.1 mg/kg SOS
  Ventilation – IPPV, $PaCO_2$ 30 mmHg, PaO2 >150 mmHg, pH >7.5
  Dobutamine 2-10 mcg/kg/m or isoprenaline 0.02 - 0.5 mcg/kg/m
  Inhaled nitric oxide: 10-80 ppm

between 6 and 18 hours after the operation, due to delayed myocardial inflammation in the process of the ischemia-reperfusion mechanism [14, 15].

The neonatal heart has a small systolic functional reserve and is much less compliant than the adult heart. To optimize its function, moderate volume loads are given (colloids: 5-10 ml/kg), together with exogenous calcium gluconate bolus (10-30 mg/kg), as the heart is both pre-load and calcium dependent. Inotropes are useful but, as the myocyte reserve is small, there is no point in insisting on extreme inotropic support. Dopamine, dobutamine and adrenaline, in small to moderate dosages, are usually enough to optimize cardiac output and adrenaline, in high dosages and for long enough, will cause myocardial necrosis. Pharmacological agents with associated vasodilator effects, such as the new generation phosphodiesterase inhibitor milrinone, are particularly useful for neonates, as they combine a mild inotropic effect with peripheral vasodilatation and afterload reduction, as well as diastolic function improvement [16]. They are named inodilators and together with dopamine (which can also have renal effects) are the first line agents after the operation. Inhaled nitric oxide (NO) is a pure pulmonary vasodilator that is useful to control pulmonary vascular tone and to improve both RV function and pulmonary gas-exchange [17, 18]. The neonatal heart has a limited stroke volume, therefore cardiac output is highly dependent on heart rate, which should be kept between 120 and 200 bpm.

Early diagnosis of cardiac dysfunction and peripheral oxygen delivery deficit is crucial in neonates. In our hands, as in others, high blood lactate levels have proved useful in predicting patients that will not survive [19], and particularly for the early detection of any cardiovascular events; lactate levels above 150 mg/dl correlated with complications, and above 200 mg/dl with mortality in the intensive treatment unit (ITU). Besides, lactate correlated well with the need for inotropes and proved to be more sensitive than pH in assessing tissue hypoperfusion and hypoxia [20]. The adequacy of surgical correction should be first verified in the event of sudden deterioration, as well as the possibility of compression or tamponade, which should always be considered. Cardiac echo at the bedside and delayed sternal closure are very useful tools to deal with these problems after the operation – if in doubt, surgical re-exploration should be promptly considered.

Renal function is very important after the operation and urine output should be kept to not less than 1 ml/kg/h. Transient low urine output periods are common and might initially be due to low cardiac output and, in some cases, to acute renal failure; measures to improve cardiovascular status are crucial, but if oliguria persists furosemide, as a perfusion – 0.5 mg/kg/m – is usually very effective. If response to furosemide is suboptimal and fluid or potassium accumulation becomes a problem, peritoneal dialysis should be started sooner rather than later, to evacuate ascites and remove both fluid and potassium. There is also some evidence that peritoneal dialysis will remove some of the inflammation mediators active after the operation [21].

Neonates that, despite well-conducted pharmacological efforts, and after a technically well conducted repair, keep on deteriorating, with persistent acidosis due to low cardiac output, or difficult gas exchange due to pulmonary hypertension or acute lung injury, should be offered the option of mechanical circulatory support, particularly by rescue extracorporeal membrane oxygenation (ECMO). This may be established electively, after difficult weaning from bypass [22], or in the ITU, as an emergency [23]. Results have improved and, currently more than 50% of post-operative patients rescued by mechanical assistance will be hospital survivors.

Table III
Managing specific cardiac lesions in the neonate

- Left sided obstructive lesions
  - Coarctation and interruption of the aortic arch
  - Critical aortic stenosis
  - **Hypoplastic left heart complex**
- Right sided obstructive lesions
  - Tetralogy of Fallot & pulmonary atresia – VSD
  - Critical pulmonary stenosis
  - Pulmonary atresia & intact septum
- Parallel circulation & mixing problems
  - Total anomalous pulmonary venous drainage
  - Truncus arteriosus
  - **Transposition of great arteries (TGA)**
    - **Intact septum – simple TGA**
    - **Complex TGA (VSD, Taussig-Bing)**

Most of the conditions that bring neonates for cardiac surgery are indicated in *Table III*, but due to space constraints we will only discuss two of the most important – **transposition of the great arteries and hypoplastic left heart complex**.

Transposition of Great Arteries

TGA is, anatomically, ventriculo-arterial discordance (pulmonary artery from the left ventricle and aorta from the right ventricle). Intact interventricular septum, patent ductus and an ASD or patent foramen ovale may be present in "simple" TGA cases; a VSD may also be present in around 20% of cases and VSD plus aortic arch obstruction occur in 10% of cases of "complex" TGA and the Taussig-Bing anomaly (double outlet right ventricle plus sub-pulmonary VSD). The coronary anatomy pattern may be abnormal, particularly in the complex forms of TGA. Diagnosis is most commonly by echo, which will identify the intracardiac anatomy as well as the coronary artery origin, but cardiac angiography may be performed, to elucidate the coronary pattern and the arch anatomy, this being done at the time of balloon atrial septostomy.

Presentation in the neonate is by cyanosis and sometimes by acidosis, due to poor blood mixing, as circulation is in parallel. Immediate resuscitation is crucial and measures to improve blood mixing are needed and include balloon septostomy and ductus re-opening by prostaglandins. In some babies, particularly in the presence of coarctation, prostaglandins are used to open the ductus and to improve mixing, but also to promote distal body perfusion, favoring diuresis and reversing acidosis.

The arterial switch operation in babies with "simple" TGA should be done electively during the first two weeks of life; patients on prostaglandins for associated coarctation or obstruction of the aortic arch are operated during the first days of life, while babies with TGA – VSD may be operated during the first three months of life, provided their clinical condition is good.

Treatment by the arterial switch operation is now standard in the neonatal period and represents 19.3% of the neonatal surgical case load registered throughout Europe, where mortality of 10% for "simple" TGA and over 15% for "complex" TGA was observed in 2002. Some aspects of surgical management still merit discussion:

• *Late presentation*

Surgical management is still a matter of discussion for "simple" TGA presenting late (beyond the first month of life), when pulmonary vascular resistance and pulmonary arter-ial pressure have dropped to normal levels. Left ventricle mass is known to involute rap-idly after birth in babies with "simple" transposition, and in the absence of a persistent large ductus, VSD or left ventricular outflow obstruction, but the LV also hypertrophies rap-idly, in response to any pressure load maintained for as little as two weeks, a capacity that is soon lost with age [24]. The question is how safe it is to offer a baby presenting beyond one month of life the switch operation, since it has been shown that operative risk increases with age, from the first week on [25]. The extreme position, accepted by very

experienced groups, is to offer the switch procedure to any baby with "simple" transposition and intact septum, presenting during the first two months of life, knowing that some will have a stormy post-operative period and will eventually need to be supported on ECMO, for some time after the operation [13]. After the first month of life it seems prudent to individualize the indications, by assessing LV mass and shape, and to offer a staged procedure – PA banding plus a 4 mm PTFE shunt, for a 7 to 10-day LV retraining period – followed immediately by the switch, whenever LV capacity is in doubt [26].

• *Surgical technique:*

The standard procedure at the moment is as follows [27]: The operation is performed through a median sternotomy, pericardium is harvested and left untreated, full mobilization of the aorta, aortic arch and ductus are achieved, followed by pulmonary artery branch mobilization, well into the lung hilum. We now conduct the operation on hypothermic cardiopulmonary bypass (18°C) and use a short period of circulatory arrest, only for ASD closure, at the beginning of the operation. The ductus is divided between ligatures, the aorta is cross-clamped and crystalloid cardioplegia (10 ml/kg) is given only once, through the aortic root. The aorta and pulmonary artery are transected above the commissures and the coronary anatomy is in-spected – dual origin is treated by excision of the coronary buttons, with a 2 mm margin of aortic tissue and little mobilization of the arter-ies, and transfer to neo-aorta openings, using the trap-door technique, to reduce tension and distortion. The Lecompte maneuver is used in virtually all cases, except where vessels are side by side, and a direct arch to neoaorta anastomosis is constructed using running monofilament suture. The author has used, over the years, a small pericardial patch anteriorly to accommodate any (aorta / pulmonary artery) diameter differences and to reduce tension on the coronary anastomosis. This procedure has proved to be both useful and safe [28].

Still with the aorta clamped, the back wall of the neo-pulmonary artery is reconstructed, using a rectangular or trouser-shaped pericardial patch and reabsorbable suture, filling in the coronary excision defects and, particularly, releasing tension from the coronary anastomosis. The anterior part of the reconstruction is performed with the heart already beating. Weaning from bypass is usually uneventful, on a small dose of dopamine and milrinone; hemostasis has to be precise. The sternum is left open, prophylactically, for 48 hours, the chest being closed with a PTFE pericardial membrane. This protocol of elective delayed sternal closure has been shown to be safe and complication-free.

In cases where the aortic arch needs to be addressed (for coarctation or interruption) – in the presence of VSD or the Taussig-Bing anomaly – one-stage correction seems to be the safer approach now [29]. In these cases the arch is addressed first and repaired during a period of circulatory arrest; however, it has been possible, over the last few years, to correct the arch anatomy by an ingenious cannulation of the innominate artery, without the need for circulatory arrest [30]. The operation then follows routinely after the arch repair.

• *Results*

Results of the switch operation have improved over the years, but the excellent figures produced by a few centers, with mortality below 1%, for "simple" TGA [13] seem difficult to reproduce, if one considers overall European figures in which operative mortality is 3.4% and 30-day mortality is 10.2% just for simple TGA [1]. Besides, results vary from center to center and mortality risk factors have long been identified [25], with results being worse with reduced center experience and the "complex" TGA and Taussig-Bing group, in which mortality ranged from 7% in selected series (31) to more than 15% (only for TGA with VSD) when unselected centers are considered as a whole [1].

Late results have been satisfactory, with good growth of the anastomosis, especially of the neo-aorta and of the coronaries, although a few patients will show some dilatation of the aortic root and aortic incompetence (around 10%, especially if a previous band had been on). Dilatation of the neo-aorta is more suggestive of normal growth, as expected for a normal pulmonary artery, than of pathological enlargement, as was shown by Karl in an elegant comparative study in 62 patients [13]. Problems related to supravalvular pulmonary stenosis are now rare and usually related to tension on the anastomosis, or to failure to use fresh pericardium for RVOT reconstruction.

Some patients will also show silent perfusion defects on myocardium scintigraphy, independent of contractility changes at rest or even on exercise, which do not seem to affect physical performance or survival [32].

Finally, neurocognitive development has been studied in a group of babies after neonatal switch in Boston, for whom circulatory arrest or low flow were used. The study showed that at least 25% of the babies suffered acutely from either seizures or EEG changes [33] and showed subnormal neurocognitive development after the operation at one and four years of age [3]. This has led us, like many others, to change our bypass policy from circulatory arrest to hypothermic full flow bypass.

Hypoplastic left heart complex (HLHC)

HLHC represents a wide spectrum of defects, comprising around 6.2% of all registered neonatal surgical cases [1]. Typically these neonates present acutely with a combination of restricted systemic blood flow (outflow obstruction, at any level) and excessive pulmonary blood flow. Anatomy is varied but the left side of the heart is smallish to hypoplastic, according to Leung's definition criteria [34] of ascending aorta < 5 mm, mitral valve < 9 mm, and LV inlet < 2.5 mm). An ASD and ductal patency are critical for survival and coarctation is commonly present. Diagnosis is usually pre-natal but will be easily done after birth; cardiac catheterization is now used only exceptionally, and may even be considered detrimental. Defining the anatomy, and also assessing right ventricular function and excluding significant tricuspid regurgitation, are essential to diagnosis.

After the diagnosis is established the baby should be kept on i.v. prostaglandins, to keep the ductus open, and be allowed to breathe room air with no added oxygen, in order to limit lung flow, by maintaining lung vascular tone. Resuscitative measures, inotropes and ventilation should be on immediate stand-by, but only used if really needed, as babies should be operated electively, in good general condition, and never as a critical emergency.

Physiological balance seems very important here, as circulation is in parallel and the objectives are to balance pulmonary/systemic resistance and flows and to support the function of the single ventricle [35]. Controlling pulmonary vascular resistance is paramount, as lung overflow will lead to systemic hypoperfusion and metabolic acidosis. Monitoring should include arterial saturation and mixed venous saturation, ideally in the range of 75% and 55% respectively, and blood lactate, both being very sensitive indicators of ideal oxygen delivery.

The possibilities for HLHC are neonatal cardiac transplantation, which is not a realistic option, due to the lack of suitable donors (36), and the Norwood staged procedure. As designed by Norwood [37], the objectives of surgery are to create an unobstructed pathway between the right ventricle (systemic chamber) and the systemic circulation (performing full arch reconstruction), to relieve any inlet obstruction between the pulmonary veins and the systemic AV valve (creating an unrestrictive ASD) and to perfuse the lungs by a controllable, non-excessive blood flow source (modified BT shunt or RV to pulmonary artery shunt). All these first stage procedures will lead optimally to a Glenn shunt (second stage) at 6 months and ultimately to a full Fontan procedure (third stage) at 2-3 years of age.

• *Surgical technique*

The first stage Norwood operation is performed by median sternotomy, cannulation of the pulmonary artery and right atrium, on cardiopulmonary bypass at 18°C, using circulatory arrest for the shortest time possible, only to reconstruct the aortic arch. Repair has recently been achieved without the need for total circulatory arrest, by perfusing through the distal BT shunt [38].

During the cooling period, full mobilization of the aortic arch, PA branches, ductus and descending aorta is performed; a PTFE shunt (3.5 mm) is then connected to the innominate artery, and will later be anastomosed to the right pulmonary artery. Under cardioplegic arrest (single injection, through the arterial cannula), the ductus is divided, the arch is incised retrogradely and the pulmonary trunk is sectioned above the commissures. The aortic arch is then reconstructed with a composite patch of pulmonary homograft, taking care not to distort the aortic root and the coronary origins and not to leave any residual obstruction, allowing for future growth [39]. The author has, on a few occasions and whenever the ascending aorta is very small, used a complete homograft tube (complete section of a

PA branch) to connect the transected PA to the inferior area of the aortic arch, this makes the repair easier and does not compromise coronary geometry. Pulmonary bifurcation is closed next with a ho-mograft patch and the proximal end of the BT shunt is then completed.

Recently, another alternative has been developed for the pulmonary blood flow source, by construction, instead of the traditional BT shunt: a 5 mm PTFE conduit between the right ventricular infundibulum and the pulmonary artery bifurcation (Sano modification – cited in [51]). This modification seems to have several advantages: more uniform pulmonary flow distribution, less pulmonary artery distortion, and particularly easier postoperative care, as pulmonary flow is only systolic, does not create aortic run-off and myocardial ischemia and is not mainly dependent on fluctuations in pulmonary vascular resistance.

Weaning off perfusion is tried on a combination of dopamine and milrinone, aiming at arterial saturations of 75% and systolic arterial pressures of 50-60 mmHg; the sternum is left open for 48 hours.

• *Post-operative care*

The critical aspect of the Norwood procedure is the postoperative period [40], as both circulations will be supported in parallel by a right ventricle ejecting through a BT shunt into the lungs and through the aortic arch to the general circulation. Balancing the flows is crit-ical: complete blood mixing should be achieved at atrial level, but resistance through the systemic and pulmonary networks will dictate the relative flows: too much pulmonary flow (due to an excessive BT shunt or to inappropriate pulmonary vasodilatation) will produce high oxygen saturations, but will create coronary run-off, reduce systemic flow and generate ominous acidosis and high lactate levels. The much better tolerated lung hypoperfusion will lead to high systemic flow rates but, if crit-ical, as when the BT shunt thromboses, will cause unacceptably low saturations and acidosis, prompting urgent shunt revision.

Monitoring after the operation should include arterial and "mixed" venous saturations, as the A-V oxygen difference correlates best with oxygen delivery, and blood lactate levels, which indicate the level of tissue perfusion [41].

Table IV

Postoperative protocol for the Norwood procedure

- **Monitoring**
  standard + *blood lactate* + Venous SAT (55%) + Art SAT (≅75%)
- **Ventilation**
  IPPV – FiO_2 21% Tidal volume 20 ml/kg to keep PCO2 40 mmHg
- **Sedation**
  Fentanyl 10 – 20 mcg/kg/h + pancuronium 0.1 mg/kg
- **Inotropes**
  Dopamine – 2-10 mcg/kg/m + MILRINONE 50 mcg/kg load + 0.25-1 mcg/kg/m
  Dobutamine – 2 to 10 mcg/kg/m
- **Hypocoagulation**
  Heparin 10 units/kg/h (try not to give platelets)

Typical Post-op Problems

- Arterial SAT >80%, increased A-V difference, raising blood lactate
  - exclude aortic arch obstruction
  - exclude myocardial ischemia & dysfunction
  - increase inotropes, increase PVR (high CO_2, low FiO_2)
  - excessive shunt – shunt revision
  - ECMO?
- Arterial SAT <40%, A-V difference constant
  - normal lactate: decrease PVR (pH>7.5, high FiO_2, low CO_2); *NO*
  - raising lactate: Heparin + shunt revision
- Normal A-V difference & SATs, but rising lactates: risk of sudden death – ECMO?

Different scenarios are possible in the failing Norwood, as indicated in *Table IV*, and immediate action has to be taken if survival is to be expected.

• *Results*

The results with the Norwood operation for hypoplastic left heart complex, as for other forms of univentricular heart, have improved over the last ten years, with selected centers producing excellent figures, with a mortality of 20% (13). Overall European figures still show operative mortality of 31.7% and in-hospital mortality of 45.1% (1). This high initial mortality will be increased by deaths related to the following stages in preparation for the Fontan procedure, making survival, at the end of the third stage, unlikely for more than 50% of all patients. Some of these patients, at any stage, may be shifted to the transplantation option. Results with cardiac transplantation for children operated below one year of age are fair, with a survival of 79.3% at one month, 69.3% at one year and 60% at 5 years [36].

Currently, the Norwood procedure is being applied to different anatomical forms of univentricular heart, some with sub-aortic

obstruction. Mortality has been related to different pre-operative factors, such as anatomic diagnosis, aortic atresia, additional defects, low birth weight and prematurity (<2.5 kg) and genetic syndromes, among others [42-49].

Sudden death during the first year after the first stage Norwood still occurs, at the rate of 4% [50] – residual lesions, coronary insufficiency, arrhythmias and shunt occlusion are all possible causes, but systemic outflow tract obstruction may also be implicated. A recent single institution study evaluated 158 patients who underwent the Norwood procedure for different forms of univentricular heart and revealed similar mortality rates for babies with hypoplastic left heart variants and other forms of univentricular hearts with associated systemic outflow obstruction. This study was unable to show any differences in mortality related to anatomy, but did demonstrate that the presence of additional cardiac or extra-cardiac anomalies was a predictor of poor outcome [51].

CONCLUSIONS

Neonatal cardiac surgery has experienced a tremendous development over the last 10 years, total correction becoming possible today for virtually any defect, from simple tetralogy to transposition of great arteries and truncus arteriosus.

Some defects are still difficult to deal with, as are complex single ventricles, for which results have improved recently.

Technical refinements in surgery, improvements in both perfusion and anesthesia, notably neurological protection, and the specialized care of dedicated neonatal units have all contributed to this level of excellence, and to the fact that the focus has now shifted more to the normal development of these future adults, to their quality of life and to their long-term survival.

In neonatal cardiac surgery, as in many other areas in life, if we wish to go much further we must step back, back into the maternal womb, to act on the fetus, possibly by modifying its faulty hemodynamics, which fetal echocardiography can now identify so early, and to promote normal great vessel and ventricle chamber growth. By doing so, how many single ventricle defects (still the major problem we face nowadays) would be gained for biventricular corrections? The future will show.

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Address for reprints:
Pedidos de separatas para:

JOSÉ FRAGATA
Department Cardiothoracic Surgery
Pediatric Unit
Hospital de Santa Marta
Rua de Santa Marta
1169-024 LISBON, PORTUGAL